

Long-Term Outcome Following Catheter Valvotomy for Pulmonary Atresia With Intact Ventricular Septum

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Objectives	This study investigated the outcome for all patients undergoing catheter valve perforation for pulmonary atresia with intact ventricular septum (PAIVS) 21 years after the first procedure at their center.
Background	Catheter perforation for PAIVS is now an established procedure. However, the management of the borderline right ventricle (RV) is controversial, and there may be a place for novel techniques such as stenting of the arterial duct.
Methods	There were 37 successful valve perforations (total 39 patients). Median length of follow-up was 9.2 years (range 2.2 to 21.0 years). Seventeen patients had stenting of the arterial duct. The mean (SD) initial z-score for the tricuspid valve was $-5.1 (\pm 3.4)$, and a further 142 sets of measurements were taken to assess the growth of the RV of survivors.
Results	There were 8 deaths (21%), and no deaths after the first 35 days. There were no late arrhythmias or ischemic events. Twenty-five patients (83% of survivors) have a biventricular circulation. For patients who had stenting of the arterial duct, significant reductions in early reintervention (0 vs. 7 patients, $p = 0.009$) and hospital stay (17.4 ± 18.1 days vs. 33.8 ± 28.6 days, $p = 0.012$) occurred, with no increase in mortality or morbidity. There was no catch-up growth of the RV in patients who had a biventricular outcome (z-score increase $+0.08/\text{year}$, $p = 0.26$).
Conclusions	Long-term survival is good, and even small RVs may be amenable to this procedure. Multiple interventions may be required to achieve biventricular circulation, but stenting of the arterial duct may reduce hospital stay and repeat procedures. (J Am Coll Cardiol 2012;59:1468–76) © 2012 by the American College of Cardiology Foundation

Pulmonary valve atresia with intact ventricular septum (PAIVS) is a rare but heterogeneous condition with an incidence of 3 to 8 per 100,000 births (1,2), and the variability with this malformation necessitates a complex decision-making process. Selection of the correct treatment pathway for patients is critical for survival (3), and there is a high early mortality without intervention (4).

The decision on the treatment pathway is based on the morphology of the right ventricle (RV) and the presence or absence of associated abnormalities (5). In many cases, treatment of PAIVS is by establishing antegrade flow

through the RV and across the pulmonary valve, with the aim of optimizing the chance of a biventricular circulation. However, some factors, such as right ventricular-dependent coronary circulation (RVDCC), are contraindications to decompression of the RV (6–9), and so a univentricular pathway is pursued.

Historically, surgery has been used in the neonatal period to establish forward flow across the pulmonary valve (PV). As an alternative, our institution performed the first catheter pulmonary valvotomy in the world for PAIVS in 1990 (10,11), and the technique is now the mainstay of treatment in many centers for patients with favorable anatomy (12–20). The technique has evolved, and stenting of the arterial duct has been added as a source of additional pulmonary blood flow in selected patients while RV function is being established (21–28). However, there remains a lack of consensus regarding the best treatment pathway for those patients with borderline anatomy (15,29).

This study reports on the long-term outcome for patients treated at our institution, where an aggressive approach has been adopted. All suitable patients, without any contraindications

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ing lesions, were put forward for catheter valvotomy. More recently, our institution has also favored simultaneous stenting of the arterial duct in selected cases. Over the long period of follow-up, the outcomes analyzed include the final type of circulation, the evolution and growth of the RV, and the safety and efficacy of stenting of the arterial duct. The study also aims to evaluate the outcomes of early decompression of the RV in those patients judged to have borderline anatomy.

Methods

Patients. From May 1990 to March 2009, 56 patients presenting with PAIVS were identified. All had simple PAIVS (situs solitus, levocardia, concordant atrioventricular and ventriculoarterial connections with confluent pulmonary arteries); 39 were put forward for catheter valvotomy. The remainder either did not survive the early neonatal period ($n = 2$), or were treated surgically with a view to creating a univentricular circulation. The criteria for surgery included RVDCC ($n = 7$), severe hypoplasia of the RV (unipartite RV) or muscular atresia ($n = 5$), or a severely dysplastic tricuspid valve (TV) or severe Ebstein's anomaly ($n = 3$).

Final outcome was evaluated on June 1, 2011. Median time elapsed since procedure was 9.2 years (range 2.2 to 21.0 years). No patient with <2 years follow-up was evaluated.

Of the 39 patients put forward for catheter valvotomy, in 2 there was failure to perforate the PV: 1 died prior to perforation due to tamponade, and the other had surgery. The baseline characteristics are outlined in Table 1.

There were 22 males and 17 females, with mean gestation of 38.1 weeks (28 to 42 weeks) and mean weight at time of procedure of 2.97 kg (0.86 to 4.47 kg). The mean z -score (30) of the TV annulus at birth was $-5.1 (\pm 3.4)$. RV to coronary artery connections were identified in 5 patients, but these were considered not to be an RVDCC. Two had mild Ebstein's anomaly, and 1 had Turner's syndrome with a bicuspid aortic valve. There were no other significant associated abnormalities.

The median age at the procedure was 4 days (1 to 72 days), with 5 of the early patients undergoing laser perforation of the atretic valve. All the others underwent radiofrequency valve perforation. The arterial duct was stented at the initial procedure in

14 patients, and at a later procedure in 3 further patients. One patient underwent a catheter pulmonary valvotomy antenatally at 23 weeks, using a 4×8 -mm Tyshak balloon (NuMED, Hopkinton, New York), with an initial good result. The membranous atresia of the PV recurred 5 weeks later, and a postnatal intervention was performed at 9 days. She currently has a biventricular circulation.

Data collection. The pediatric cardiology database of the Evelina Children's Hospital and Guy's Hospital was reviewed retrospectively to identify all patients with PAIVS. The first catheter valvotomy procedure was performed in May 1990, and a minimum of 2 years follow-up was required for inclusion. Final outcomes are correct as of June 1, 2011.

Data for those patients, who had been put forward for catheter valvotomy, were gathered from the clinical notes and hospital database, including baseline demographics. The clinical course for each patient was recorded, including significant complications (intra-procedural, vascular, sepsis, neurological, other), length of intensive care unit and in-patient stay, further catheter and surgical procedures, and final outcome in terms of circulation type and disability. Longitudinal data were also collected when recorded, including weight, height, oxygen saturation levels, and valvar dimensions. In total, 142 sets of measurements were collected.

Ethical approval was granted by Guy's and St Thomas's Ethical Committee and Research and Development Committee (Reference RJ111/N028).

Baseline echocardiographic dimensions. The initial echocardiogram after birth was reviewed for a representative sample of 18 of the 39 patients, in order to assess reproducibility of baseline data. The echocardiograms were measured independently by 3 investigators (H.C., J.M.S., and S.A.Q.) on 2 separate occasions, and the mean of these values was recorded. The tricuspid and mitral valves were measured from the apical 4-chamber view in early diastole (Fig. 1), the aortic valve in a long-axis parasternal view and the PV in a short-axis parasternal view in early systole. Measurements were performed offline using electronic calipers (Hewlett-Packard Sonos 7500, Philips, Andover, Massachusetts) for echocardiograms performed before 2007, and using EchoPAC software package (GE Medical Systems, Milwaukee, Wisconsin) for echocardiograms performed since 2007. The z -scores were calculated from the algorithms established by Daubeney et al. (30). Using the averaged values of the 3 investigators as a gold standard, the measurements were compared with those documented in the clinical records contemporaneously. A Bland-Altman analysis (31) was utilized and the agreement was assessed as acceptable (mean bias of [averaged reviewed values] –

Abbreviations and Acronyms

- MV** = mitral valve
- NEC** = necrotizing enterocolitis
- PAIVS** = pulmonary atresia with intact ventricular septum
- PV** = pulmonary valve
- RV** = right ventricle
- RVDCC** = right ventricle-dependent coronary circulation
- TV** = tricuspid valve

Table 1 Summary of Baseline Characteristics of Population for Catheter Valvotomy ($N = 39$)

Birthweight, kg	2.97 \pm 0.57
Gestational age, weeks	38.1 \pm 3.4
Antenatal diagnosis	20 (51)
Sex, male	22 (56)
Age at initial procedure, days	6.9 \pm 11
TV annulus size, cm	0.94 \pm 0.25
TV annulus z -score	-5.1 ± 3.4
TV/MV annulus ratio	0.71 \pm 0.22
Mean length of follow-up, yrs	10.7 \pm 6.1

Values are mean \pm SD or n (%). Annulus measurements were obtained from the apical 4-chamber view.

MV = mitral valve; TV = tricuspid valve.

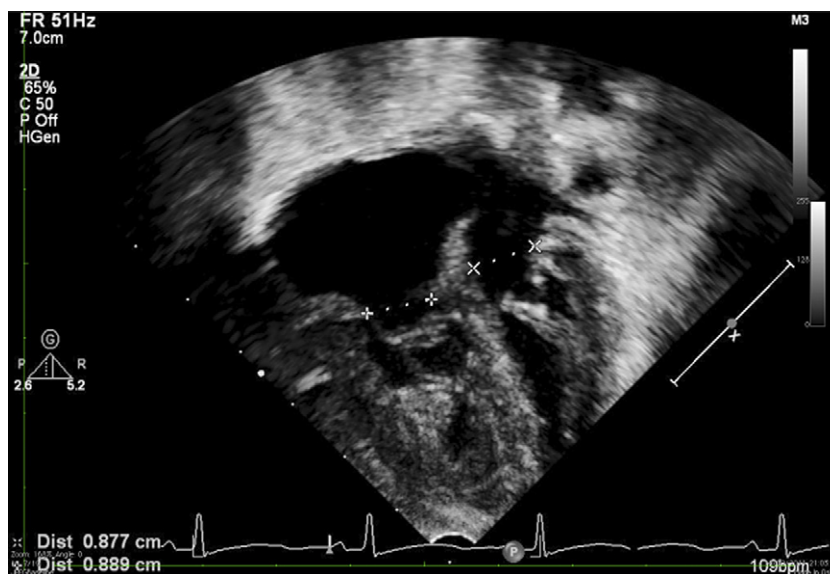


Figure 1 Echocardiogram Showing 4-Chamber Apical View in Early Diastole

The measurements of the tricuspid and mitral valves were taken from the inner septal and lateral wall margins of the annulus. Measurements were taken by 3 independent observers (H.C., J.M.S., and S.A.Q.) on 2 separate occasions and averaged.

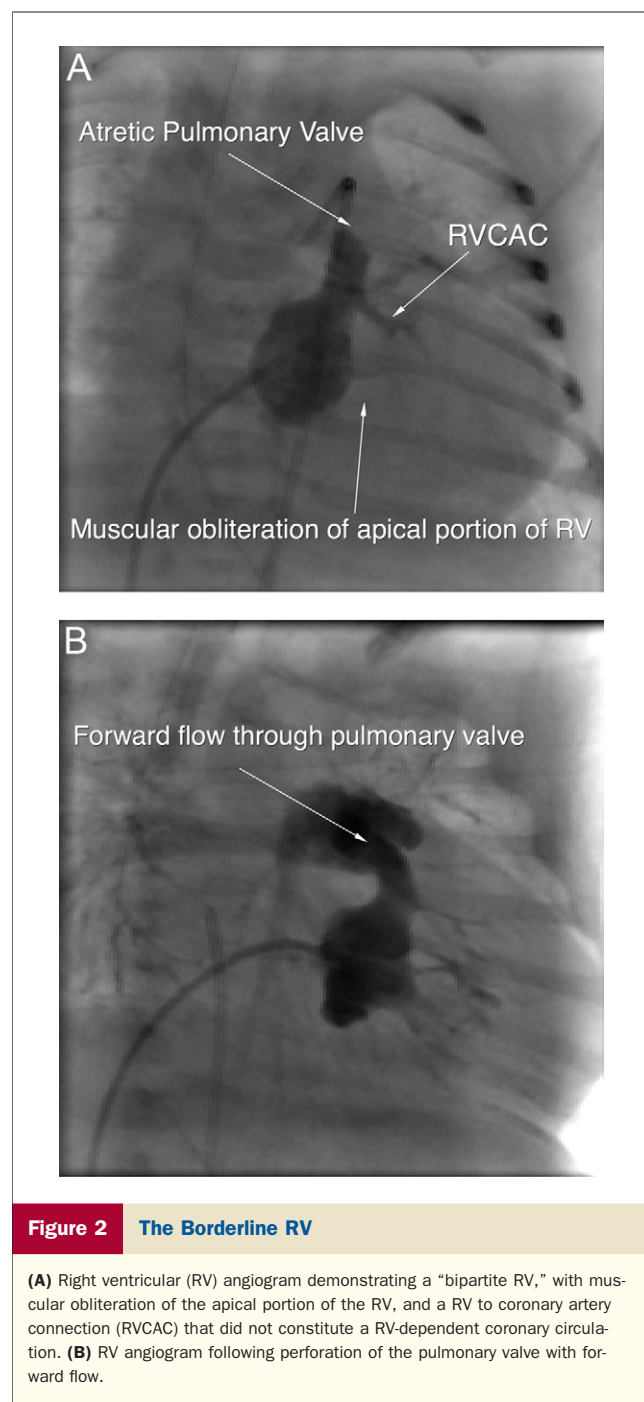
[clinical record values] + 0.09 cm, SD: 0.15 cm), and the measurements recorded in the patient notes at the time of initial assessment were accepted for the rest of the cohort. Longitudinal measurements over time of the valve annulus of each survivor were also made, through review of the original echocardiogram. Where this was not possible, contemporaneous measurements were accepted. Using the weight and height for the child recorded at the time of the echocardiogram, the valve z-score for each point in time was calculated.

Procedure. The technique of the catheter procedure has been described in detail in earlier publications from our center (10,12,22,32). Briefly, cardiac catheterization was performed under general anesthesia in all neonates with full heparinization and antibiotic cover. Vascular access of the femoral vein and femoral artery was obtained percutaneously, and 4-F sheaths and 4-F catheters inserted. Initial angiography of the RV body and outflow tract was performed in order to determine the size and morphology of the RV and the presence or absence of sinusoidal connections from the RV to the coronary arteries (Figs. 2A and 2B). Catheters were positioned through the femoral venous and arterial routes just below and above the atretic PV, respectively. Once stable and acceptable positions of the catheters were obtained, a laser (argon generator, “hot tip” wire) or radiofrequency wire (5 W for 2 to 5 s was used, although occasionally 10 W were required) was advanced through the venous catheter and the heated tip gently pushed to perforate the atretic valve. Using a gooseneck snare and a super-floppy guidewire, an arteriovenous circuit was established in the majority of cases to allow the safe passage of

the appropriately sized low-profile Tyshak balloon across the valve. The PV was dilated with a balloon to 100% to 120% of its size.

Simultaneous stenting of the arterial duct was performed in selected patients from 2000. The duct was stented if it was judged by the interventionalist that the patient was likely to require prostaglandin therapy for more than 2 weeks. The judgment was subjective, and was based upon the size and morphology of the duct and perceived adequacy of the RV size and function. If the decision had been made to stent the arterial duct, the prostaglandin infusion was stopped at the beginning of the procedure. The valve was perforated and ballooned as described in the previous text, then the duct was assessed angiographically and the stent size selected (Figs. 3A to 3D). The stent was generally positioned across the duct from the aortic side, with multiple handshot angiograms in order to confirm good stent position before inflation.

Statistical analysis. Statistical analyses were performed using Stata v. 11 (StataCorp, College Station, Texas). Summary statistics are expressed as mean (\pm SD) for normally distributed variables, and as median (range) for non-normally distributed variables. Two sample comparisons utilized either the *t* test or Wilcoxon rank sum (Mann-Whitney) test dependent upon normality of the variable’s distribution. Fisher exact test was employed for binary variables. Growth trajectories for the TV annulus were modeled using a random coefficients model (xtmixed command), and comparisons of TV growth trajectories were made between patients who attained biventricular versus univentricular circulations.



Results

Catheter valvotomy was successful in 37 of 39 patients, defined by the completion of valve perforation and balloon dilation of the RV outflow tract. In 1, the procedure was abandoned due to unsatisfactory positioning of the catheters for attempted valvotomy (TV z-score: -11.3). The other patient (weight at procedure 860 g, TV z-score: -16.4) died in the catheter laboratory following cardiac tamponade due to guidewire perforation of the RV outflow tract.

Hemodynamics. Prior to perforation of the PV, the mean RV systolic pressure was $101 (\pm 28)$ mm Hg, and the mean aortic systolic pressure $68 (\pm 17)$ mm Hg. The mean pulmonary artery systolic pressure prior to perforation was $37 (\pm 15)$ mm Hg. Following perforation and dilation of the PV, the systolic RV, pulmonary artery, and aortic pressures were $51 (\pm 20)$ mm Hg, $36 (\pm 9)$ mm Hg, and $63 (\pm 12)$ mm Hg, respectively. The median size of the balloon used to dilate the PV was 7 mm (range 5 to 10 mm).

Mortality. There were 8 deaths (21%) in total. One patient (described in the previous text) died at surgery following an unsuccessful valve perforation. Of the other 7 deaths, 2 occurred due to cardiac tamponade, 3 due to sepsis (including 2 with necrotizing enterocolitis [NEC]), and 1 following a cerebral hemorrhage at 22 days. One occurred after the patient developed profound hypotension, resistant to inotropes, and the impression at post-mortem examination was that this was most likely attributable to a “figure of 8 circulation,” with a stented arterial duct and regurgitant PV and TV.

Complications. Cardiac tamponade occurred in 6 patients during the procedure, including those 2 who later died. In 1 patient, the ductal stent embolized to the aorta and was successfully retrieved. There were 9 who required thrombolysis for femoral vascular occlusion following the procedure, and all but 2 recovered normal pulses.

Significant long-term comorbidities following the procedure were neurological deficits in 2 patients. Both had a hemiparesis, 1 due to a thalamic hemorrhage (day 1 after procedure, following thrombolysis with streptokinase) and 1 due to a cerebral abscess (diagnosed day 92 after the procedure). One had cerebral palsy, but this was thought to be due to an undiagnosed syndrome. One has suffered from seizures that were controlled on medication.

Long-term outcome. The 30-day mortality for all 39 attempted procedures was 15% (6 deaths). Of the 37 successful procedures, there were 7 (19%) deaths, all occurring within the first 35 days post-procedure. Six occurred during the first 10 years of experience, and 2 in the subsequent 10 years. The causes of death are as detailed above, and there were no late deaths. There were no late arrhythmia or ischemic events. Figure 4 summarizes the final outcome for the population.

At the most recent follow-up, 25 (83%) of the 30 survivors had a biventricular circulation. Eighteen patients with a biventricular circulation required a further procedure, either surgical or catheter based (Fig. 5). Repeat balloon dilation of the PV was required in 13, and 6 required balloon dilation or late stenting of the arterial duct. Insertion of a Blalock-Taussig shunt was required in only 3 patients. Two achieved biventricular circulation but needed a surgical right ventricular overhaul, at 25 and 39 months, and this technique has been described in detail in other publications (29,33); 1 required a surgical reconstruction of the RV outflow tract at 6 months. Twelve required closure of an atrial septal defect. A device occlusion was performed in 5, at a mean age of $6.6 (\pm 2.7)$ years. The remaining 7 had

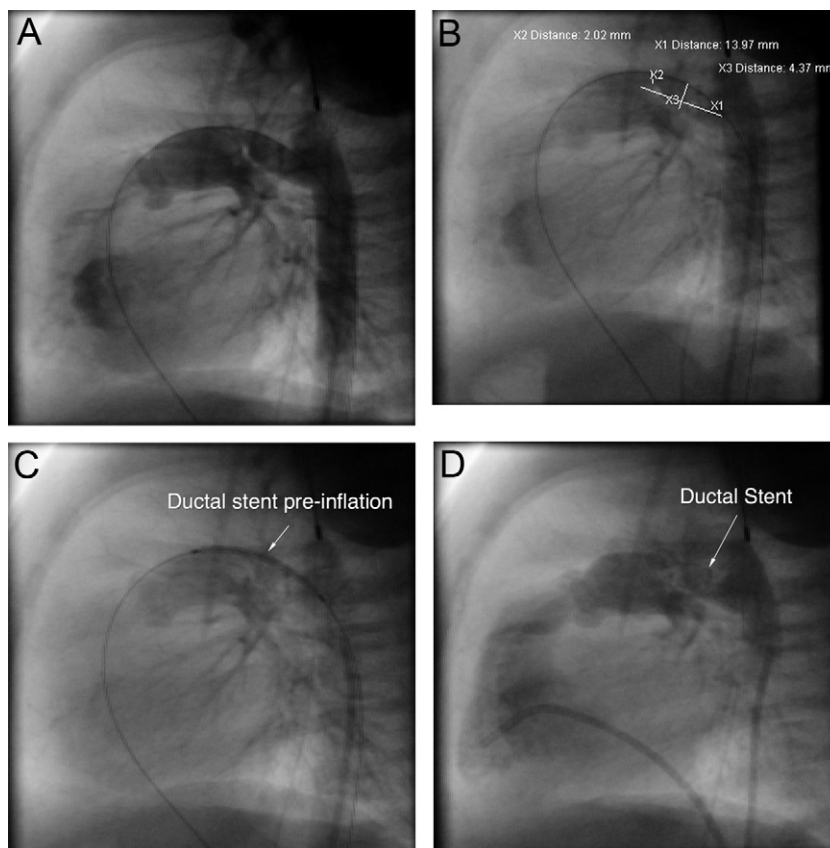


Figure 3 Stenting of the Arterial Duct

(A and B) Aortic angiogram demonstrating the morphology of the arterial duct and subsequent measurements. Note the secure wire position across the arterial duct. (C) Ductal stent in position, inserted from aortic side, prior to inflation. (D) Aortic angiogram demonstrating flow through the ductal stent. Note the regurgitation into the right ventricle through the perforated pulmonary valve.

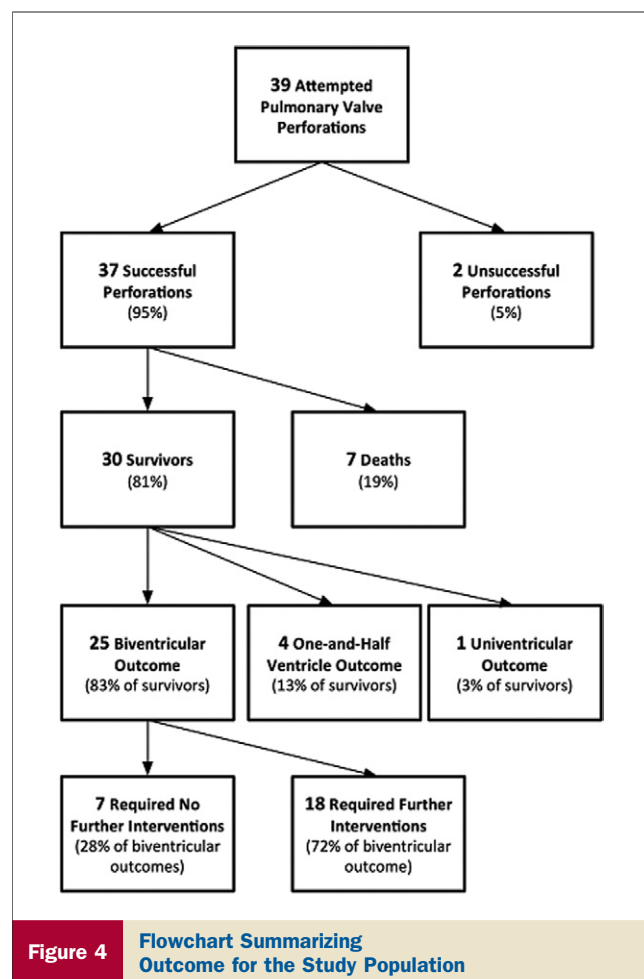
surgical closure, 4 in conjunction with another procedure, at a mean age of 2.5 (± 2.0) years.

Of the 5 (13% of all procedures) patients not achieving biventricular circulation, 4 have a 1-and-a-half ventricle circulation completed at a mean of 23 (± 15) months, and 1 has a Fontan circulation, completed at 49 months.

Stenting of the arterial duct. The arterial duct was stented at the initial procedure in 14 patients, and at a subsequent procedure in a further 3 (at 11, 27, and 44 days post-procedure, respectively). Four further patients in the early years of the technique had ballooning of the arterial duct alone. In 1, stenting of the duct was attempted at the initial procedure, but the stent embolized to the descending aorta and was retrieved successfully. The decision was made not to stent the duct of this patient. There were 2 deaths in the cohort that were stented, 1 of which may be attributable to the stenting of the duct, with a “figure of 8 circulation” as described previously. The other death was due to klebsiella sepsis. Table 2 summarizes the characteristics and outcomes of patients, divided according to whether the arterial duct was stented.

The length of stay in pediatric intensive care and in hospital was significantly shorter for those in whom the arterial duct was stented, and early reintervention was significantly reduced. There was no increase in mortality or incidence of NEC in the stented group. In the long term, 1 patient required redilation of the ductal stent at 12 months, and this was performed at the same procedure as balloon dilatation of the RV outflow tract. No patient required the insertion of further ductal stents at a later procedure. All but 1 ductal stent occluded spontaneously following cessation of aspirin. One patient required occlusion of the ductal stent with a coil performed at 40 months at the same procedure as a device closure of the atrial septal defect. There was no report of late branch pulmonary artery stenosis.

Predictors of final outcome. Patients with a smaller initial TV annulus and PV annulus z -scores were less likely to achieve biventricular circulation and more likely to have an unsuccessful procedure/death (Table 3). For patients with a reduced TV/mitral valve ratio, there was also a trend towards a non-biventricular outcome, but this was not significant. Higher saturations ($>90\%$) at 1 year were highly correlated with an eventual biventricular outcome. Those



with an antenatal diagnosis were more likely to achieve a biventricular outcome. There was no relationship between birthweight or gestation and outcome.

Growth trajectories. TV growth trajectories via the random coefficients model are shown in Figure 6. Patients achieving biventricular outcome demonstrated higher TV z -scores at baseline (-3.92 vs. -7.01 , $p < 0.001$). Those achieving a univentricular circulation demonstrated a decrease in TV z -score with time (approximating -0.38 units per year, $p < 0.001$). Conversely, patients achieving biventricular circulation showed a trend towards an increase in TV size and z -score with time. However, this was not sufficient to constitute “catch-up” growth, as the z -score did not change significantly (gradient $+0.08$ per year, $p = 0.26$).

Discussion

This study represents the longest follow-up to date of a population with PAIVS who have undergone catheter pulmonary valvotomy (14,16–18). However, this population also represents a departure from management algorithms generally used in other studies. This difference is in treatment of the borderline RV and the role of stenting of the arterial duct at the initial procedure.

The borderline RV. It is important to choose the correct treatment pathway, balancing the need for short-term safety against the long-term goal of maximizing quality of life. Ashburn et al. (3) demonstrated the importance of balancing the desire for a biventricular repair against the cardiac anatomy, and that centers that strive too hard to achieve an ambitious repair run the risk of high mortality in their surgical patient population. This principle should also apply to catheter valvotomy procedures.

However, decompressing the RV, unless contraindicated by anatomical features such as RVDCC, will be advantageous in the long term regardless of the final outcome (29,34). The advantages include improved left ventricular function (16), growth of the RV (34–36), reduction in endocardial fibroelastosis, and stimulation of development of the pulmonary vasculature (37). There are additional benefits for the patients if these can be achieved by catheter intervention rather than surgery (38).

If the likelihood of achieving biventricular circulation is not an essential criterion for catheter valvotomy, the question still arises as to what criteria should be used to identify an RV as being unsuitable for the procedure. A number of cutoff points have been suggested in other publications, including a TV annulus of at least 11 mm (19) or a z -score of at least -3.5 (14,17). In our study, these values appear extremely conservative, but it is important to bear in mind the algorithm with which a z -score is calculated. Our study has chosen to use the algorithms calculated from the data of Daubeney et al. (30), in common with the majority of comparable publications (5,14,16,34,36). However, some studies have used algorithms calculated from autopsy measurements (18,39) or have not made explicit which set of algorithms have been used. This is relevant, as the z -score of the TV can vary by as much as 3 standard deviations between different algorithms.

Over the 21 years since the first procedure, our center has maintained an aggressive approach towards catheter valvotomy. Patient selection has remained unchanged, and all patients without any contraindicating features have been put forward for the procedure. This is despite the relatively high mortality rate in the early era. The procedure has evolved to become an accepted and relatively safe treatment strategy in many centers.

Stenting of the arterial duct. The arterial duct was stented in 14 patients at the initial procedure, including the majority (85%) from 2004 onwards. This approach represents a departure from that advocated in other publications, although a recent paper from Alwi et al. (27) has also demonstrated promising results following stenting at the initial procedure. In patients in whom the arterial duct was stented, there was a significantly reduced re-intervention rate and hospital stay.

The most significant complication is the development of high-flow cardiac failure in 1 patient, who died. The precise cause of the hemodynamic instability in this patient is difficult to determine as the stent used in the duct was of a

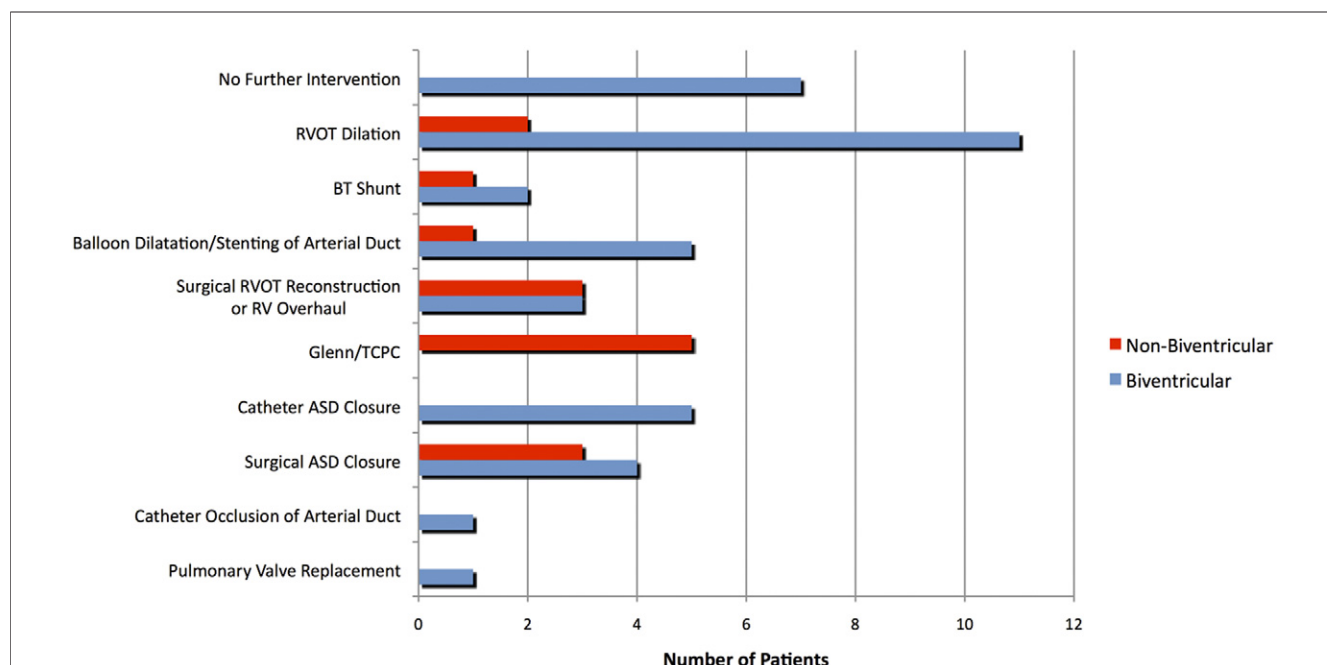


Figure 5 The Number of Subjects Who Underwent Further Procedures After the Pulmonary Valvotomy

ASD = atrial septal defect; BT = Blalock-Taussig; RV = right ventricle; RVOT = right ventricular outflow tract; TCPC = total cavopulmonary circulation.

similar size to that used in other cases with a similar birthweight (4 × 16-mm Liberte Stent, Boston Scientific, Natick, Massachusetts), and the initial post-procedure pulmonary and tricuspid regurgitation was no more severe than for most patients undergoing this procedure. Alwi et al. reported a single death in their cohort with a comparable pathology. In our cohort, there was no increased incidence of NEC, renal failure, or vascular complications. In the long term, all but 1 duct closed spontaneously following cessation

of aspirin. The final case required closure of the arterial duct with a coil, and the proportion requiring interventional closure is similar to other studies (21,26).

Growth of the RV. The TV annulus measurement was used as the surrogate for RV size in this study. Several studies in the past have failed to demonstrate catch-up growth of the TV in terms of an increase in the TV z-score over time (16,29,35,36,40), and this was replicated in this study. It is important to note that catch-up growth of the RV is not an essential criterion for establishing a biventricular circulation. In separating out those that did not achieve a biventricular circulation, cause and effect are confounded. However, for those with a non-biventricular outcome, the fall in z-score generally predates the creation of a cavopulmonary shunt. These findings are in contrast to Huang et al. (40) who found the greatest potential for catch-up growth in those with the smallest RV.

Achieving biventricular circulation and the long-term outcome. Eighty-three percent of survivors in our study currently have a biventricular circulation, similar to comparable studies (14,16–18). The improvement in survival figures over time reflects the learning curve associated with a new and technically demanding technique that continued to develop (41,42).

Following successful perforation of the PV, those with a smaller RV tend to require more interventions in their treatment pathway. This staged approach should not be seen as a failure of the initial procedure, but rather as the tailoring of treatment to fit such a heterogeneous group (26,29,43). It is

Table 2 Summary of Characteristics of Patients Who Did or Did Not Undergo Stenting of the Arterial Duct Following Successful Valve Perforation

	Stented (n = 17)	Not Stented (n = 20)	p Value
Year	2005 (1992–2009)	1998 (1990–2009)	0.003
zTV	−5.11 ± 2.53	−4.13 ± 2.92	0.28
Days ICU	4.6 ± 2.9	9.1 ± 8.0	0.029
Days in hospital	17.4 ± 18.1	33.8 ± 28.6	0.012
NEC	2 (12)	6 (30)	0.25
Early reintervention	0	7 (35)	0.009
Late reintervention	11 (65)	8 (40)	0.13
Biventricular outcome	11 (65)	14 (70)	0.92
Death	2 (12)	5 (25)	0.32

Values are median (range), mean ± SD, or n (%). p Values in bold are statistically significant. zTV indicates the z-score of the TV annulus (Daubney algorithm [30]). Early reintervention is defined as further catheter or surgical intervention in the first 100 days following the valvotomy. Five of the reinterventions were the insertion of a Blalock-Taussig shunt. Stented group includes 3 patients who were stented at a later procedure, at 11 days, 27 days, and 44 days post-procedure, respectively, and these are not included as early reinterventions. Late reintervention is defined as further catheter or surgical intervention >100 days following the valvotomy (4 patients in the unstented group had both early and late reinterventions).

ICU = intensive care unit; NEC = necrotizing enterocolitis; TV = tricuspid valve annulus.

Table 3 Baseline Characteristics and Predictive Value for Final Outcome

	Died/Unsuccessful n = 9 (23%)	Non-Biventricular Outcome n = 5 (13%)	Biventricular Outcome n = 25 (64%)	p Value
Gestation, weeks	37.8 ± 6.4	38.5 ± 1.8	38.0 ± 2.1	0.54
Birthweight, kg	2.88 ± 0.91	2.86 ± 0.24	3.02 ± 0.48	0.50
Antenatal diagnosis	2 (22%)	2 (40%)	16 (64%)	0.048
Male	6 (67%)	3 (60%)	13 (52%)	0.52
Age at procedure, days	14.7 ± 22.6	4.6 ± 2.3	4.6 ± 3.7	0.37
Duct stented	2 (22%)	4 (80%)	8 (32%)	0.51
RV pressure before procedure	112 ± 35	89 ± 29	102 ± 26	0.98
Change in RV pressure, mm Hg	−58.4 ± 40.7	−19.2 ± 42.5	−50.6 ± 24.4	0.44
zPA	−2.76 ± 0.63	−3.91 ± 1.31	−1.97 ± 1.43	0.005
zTV	−7.36 ± 5.4	−7.01 ± 1.84	−3.92 ± 2.10	0.020
TV/MV annulus ratio	0.63 ± 0.25	0.60 ± 0.08	0.76 ± 0.22	0.06
Saturations at 1 yr, %	NA	78.5 ± 4.5	93.4 ± 4.3	0.008
Follow-up, yrs	NA	8.4 ± 1.8	10.1 ± 6.0	

Values are mean ± SD or n (%). p Values in bold are statistically significant. For the statistical analyses, the died/unsuccessful and non-biventricular outcome groups have been combined. Z-scores were calculated using the algorithm of Daubeney *et al.* (30).

RV = right ventricle; TV/MV ratio = ratio of the size of the TV annulus to the mitral valve (MV) annulus; zPA = the z-score of the pulmonary valve (PV) annulus; zTV = z-score of the tricuspid valve (TV) annulus.

also interesting that this study suggests that there may be a survival and final outcome advantage for those patients that have been diagnosed antenatally. This is in contrast to earlier studies (44) and requires further research.

Late complications are rare. Those patients with forward flow across a small TV into a restrictive RV might be expected to be more likely to develop a dilated right atrium and arrhythmias. However, to the best of our knowledge, this has not been reported, and none of the patients in this cohort has suffered from this complication. There has been concern in the literature that patients with PAIVS may go on to develop myocardial perfusion abnormalities (45), but the cohort most likely to develop this are those with RVDCC, who were excluded from this study. There was no event suggestive of myocardial hypoperfusion in either the

early or late stages of follow-up, although no patient has been formally investigated for this.

Conclusions

Over the time period of this study, the technique of catheter pulmonary valvotomy has evolved from a pioneering intervention to become a mainstay of treatment for PAIVS at many institutions. Safety continues to improve, and a balance is required between providing the RV with the best possible chance of supporting the cardiac output against the increased risk of the procedure in borderline cases. This study supports a more aggressive approach to achieving 2-ventricle circulation, with intervention in some cases with a small RV that otherwise may not have been considered for catheter valvotomy. Additionally, prophylactic stenting of the arterial duct should be considered in those who are judged most likely to require early reintervention. In the long term, mortality outside of the acute phase of treatment is extremely low, and late complications are rare.

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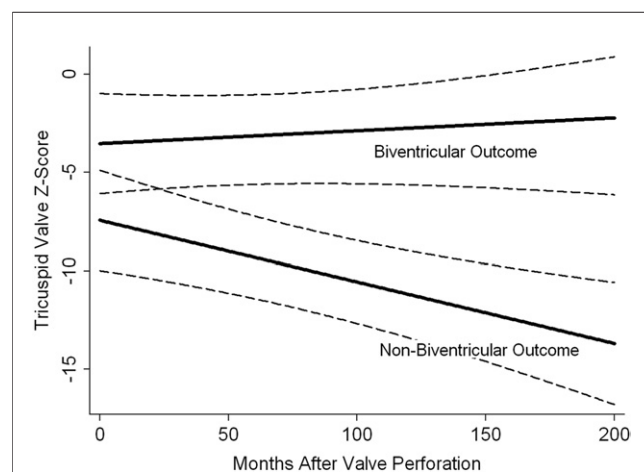


Figure 6 Calculated Growth Trajectories of the Tricuspid Valve

Z-scores are calculated using the algorithm derived by Daubeney *et al.* (30). **Solid lines** represent predicted values (from the random coefficients model); **dashed lines** indicate 95% confidence intervals.

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Key Words: arterial duct ■ biventricular circulation ■ catheter valvotomy ■ hypoplastic right ventricle ■ laser perforation ■ pulmonary atresia with intact ventricular septum ■ radiofrequency perforation ■ stent.